

AMENDMENTS TO THE SPECIFICATION

Please replace the final paragraph on page 3 (spanning lines 31-36) with the following:

Figure 4 shows the nucleic acid sequence for the $\gamma 2$ chain cDNA and the derived amino acid sequence. Figure 4A is the full cDNA for the 5,200 base pair sequence, available from EMB/GenBank/DDBJ under the accession number Z15008 (SEQ ID NO: 12 and SEQ ID NO: 13). Figure 4B is the nucleotide (SEQ ID NO: 14) and derived amino acid sequence (SEQ ID NO: 15) of the alternative 3' end sequence from cDNA clones providing a sequence of 4,316 base pairs, available from EMB/GenBank/DDBJ under the accession number Z15009. (Kallunki, et al., 1992, *supra*.)

Please replace the first two paragraphs of page 11 (spanning lines 1-16) with the following:

PCR reactions on genomic DNA (50 μ g) were carried out using the upstream primer 5'-TTCCTTTCCCCTACCTTGTG-3' (SEQ ID NO: 18) and the downstream primer 5'-TGTGGAAGCCTGGCAGACAT-3' (SEQ ID NO: 19), which are located in the intron 2 and exon 3 of LAMC2 respectively. PCR conditions were: 95 C, 5 min, followed by 94 C, 45 sec; 56 C, 45 sec; 72 C, 45 sec; for 35 cycles, and extension at 72 C for 5 min. PCR products were used for restriction analysis. 20 μ L of PCR product obtained from genomic DNA was digested with TaqI for 2 hours (Boehringer Mannheim). Cleavage products were electrophoresed (2.4 % agarose) stained and visualized under UV light.

Thus the methods allow for the screening of patients for mutations in the $\gamma 2$ chain which correlate with H-JEB. As demonstrated, the results have identified a nonsense mutation resulting in a truncated $\gamma 2$ chain, leading to severe H-JEB. This was further confirmed by specific amplification and restriction enzyme analysis of both the patient and relatives. Thus demonstrating the effective screening for and identification of, $\gamma 2$ chain mutations which correlate with H-JEB. The methods are thus useful for diagnosis, prenatal screening, early screening and detection, as well as detailed examination of H-JEB. Furthermore, the results demonstrate the significance of the $\gamma 2$ chain in forming proper cellular contacts.